

Letter to the editor

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The Raveenthiran Syndrome—Ectopic testis within Spigelian hernia: Few additional remarks

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Dear Editor

I read, with great interest, the article by Sinacer et al. on the association of congenital Spigelian hernia with cryptorchidism, polydactyly, and anal stenosis.[1] Although this association is known since 1895, only in 2005 it was recognized as a new syndrome.[2,3] Since then there has been an increasing awareness about this clinical entity.[4] In 2015 Moles-Morenilla et al. named it 'Raveenthiran syndrome' and classified it into two types: type-1 is an exclusive association of Spigelian hernia and ectopic testis while type-2 is Spigelian hernia and ectopic testis associated with other anomalies.[5] Accordingly, the newborn reported by Sinacer et al. belongs to type-2 Raveenthiran syndrome.

Pathogenesis of this syndrome is conjectural. Previously Spigelian hernia was thought to be the primary defect and ectopic testis be a sequel of that. However, I rightly deduced that ectopic testis is the primary anomaly, while Spigelian hernia is a secondary phenomenon in which raised intra-abdominal pressure converts the potential sac (processus vaginalis) accompanying ectopic testis into Spigelian hernia. Subsequently, John Hutson agreed with my theory of pathogenesis and gave a molecular explanation of it. [6] The factor that prompted my hypothesis was the association of anal stenosis. Interestingly, Sinacer's case is also associated with anal stenosis, thus adding strength to my hypothesis. Such a combination has been reported only thrice in the literature. [5]

Miliras objected to the term 'syndrome' because in dysmorphology it is reserved for the set of anomalies

that are independently caused by a known common etiological factor. [7] He suggested that 'sequence' is the correct terminology as the Spigelian hernia is a mechanical consequence of ectopic testis. The term 'syndrome' is used in two different contexts. Its clinical and dysmorphological usage should not be confused. Etymologically 'syn' means 'together' and 'dromos' means 'running' ('syndrome' means 'occurring together'). In dysmorphology, the term is used in etiological connotation while in clinical practice it is used in an etymological sense. This assertion is supported by the existence of clinical terms such as 'nephrotic syndrome' and 'MIS-C' (multisystem inflammatory syndrome of children due to Covid-19).

Finally, I disagree with the authors in postponing the treatment of anal stenosis. Since raised intra-abdominal pressure is the pathogenic precipitating factor of Spigelian hernia, priority correction of anal stenosis is essential to prevent recurrence of the hernia. I also disagree with the authors' suggestion of using a mesh to prevent a recurrence. Relocating the ectopic testis into the scrotum and anatomical repair of the abdominal wall defect using absorbable sutures is sufficient in treating Raveenthiran syndrome.

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